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EVALUATION OF SURGICAL COMPLICATIONS IN SICKLE CELL HEMOGLOBINOPATHY, AT VIMSAR,BURLA

General Surgery

Dr. Sribasta Kumar Mahapatra Professor,dept. Of General Surgery,vimsar,burla,

Dr. Rohit Gohil Asst. Prof., Dept. of General Surgery, VIMSAR, BURLA.

Dr Satya Prakash Sahoo* Junior Resident, VIMSAR, BURLA. *Corresponding Author

ABSTRACT

INTRODUCTION: Sickle cell disease is a structural abnormality of haemoglobin , with autosomal recessive inheritance . The term sickle cell is derived from sickle shaped appearance of RBC. It is due to point mutation (GAG→GTG;Glutamic acid to valine) that occurs in the sixth position of the β-globin part of haemoglobin.

AIM OF THE STUDY: To study the surgical complications and their management in patient with sickle cell haemoglobinopathy.

METHODS: We took 140 patients in dept. of general surgery ,VIMSAR,BURLA from December 2018-November 2020.It was a observational study.

DISCUSSION: The incidence of surgical complications of sickle cell disease like cholelithiasis, splenomegaly, chronic duodenal ulcer , priaprism, acute abdominal crisis ,chronic leg ulcer, were found in present series were 30%,51%,4%,2%,2% ,4%respectively.

CONCLUSION: It is hoped that the increasing awareness of the surgical manifestations of SCD, the introduction of minimally invasive technique and the new preventive therapies such as hydroxyl-urea aim at reducing the number and severity of sickle cell crisis thus reduce the high mortality and morbidity associated with surgery.

KEYWORDS

Sickle cell disease, splenomegaly, priaprism, cholecystectomy

INTRODUCTION:

Sickle cell disease is a structural abnormality of haemoglobin ,with autosomal recessive inheritance . The term sickle cell is derived from sickle shaped appearance of RBC. It is due to point mutation (GAG→GTG;Glutamic acid to valine) that occurs in the sixth position of the β-globin part of haemoglobin.

About 250 million people, i.e. 4.5% of world population are carrier and 300000 infants are born with major haemoglobinopathies every year, 70% the world wide haemoglobin disorder. As per WHO report ,60 million carriers of sickle cell gene and 1,20,000 sickle cell homozygotes are added every year in the world. It is estimated that there were 24,34,170 carriers and 1,21,375 sickle cell homozygotes among the tribes of india.and 3-4 million people are suffering from sickle cell hemoglobinopathies in odisha alone.

Comparision to world and national figures, the prevalance of hemoglobinopathies in odisha about 19.32%,out of which 13.2% suffer from sickle cell hemoglobinopathies.(sickle cell trait=8%,sickle cell disease=4%,sickle cell beta thalasemia =1.2%).

Two phenomena namely ,sickling of cell with occlusion of vessels and hemolysis of cells are responsible for clinical manifestations of disease.

METHODOLOGY:

Place of study: Dept. Of general surgery,VIMSAR,Burla
Period of study: From December 2018-November 2020
Study design: Crosssectional observation study
Study population: All in patient ,who have sickle cell disesase with surgical complications in all surgical units of VIMSAR.
Sample size:140

INCLUSION CRITERIA:

1. Patients, presented with sickle cell disease with surgical complications.
2. Patients,who gave consent.

EXCLUSION CRITERIA:

1. SCD with skeletal involvement
2. SCD with renal involvement.

DISCUSSION:

The present study comprised of 140 cases of sickle cell disease admitted to General Surgery wards of V.S.S. Medical College, Burla from December 2018 to November 2020.

The observations and discussions in this thesis works dealt with :-

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

Showing Age Incidence

Age group in years	No. of cases	Percentage
1-10	29	20.71%
11-20	59	42.14%
21-30	38	27.14%
31-40	12	8.57%
41-50	2	1.42%
>50	0	0
Total	140	100%

The table above, it is that the incidence of sickle cell disease with surgical complications is highest in the age group of 11 to 20 and again from 21 to 30 years and the total being 69.28% (42.14% + 27.14%)

The lowest age of the case studied here is 5 years and the highest is 45 years. As the age advances beyond 30 years, which is most active phase of life, the incidence of surgical complications of sickle cell disease gradually declines.

TABLE
Showing Sex Incidence

Sex	No. of cases	Percentage
Male	86	61.42%
Female	54	38.57
Total	140	100

From the above table it is evident that the male members with sickle cell disease present with surgical problem in 61.42% of cases as compared to females accounting for rest 38.57% with a male female ratio of 8:5.

However the sex incidence so far as the surgical complications are concerned has not been studied in detail by any previous workers. But male and female members present different surgical problems, which are confirmed to the particular sex like priapism in males and secondary amenorrhoea or problems of pregnancy in females.

Table Incidence Of Surgical Complications In Different States Of Sickle Cell Disease

State of sickle cell disease	No. of cases	Percentage
Steady state	137	97.85%
Crisis	3	2.14%
Total	140	100%

The above table depicts that, out of 140 cases, only 3 numbers of cases presented with sickle cell crisis. The rest, though presented in a steady state, might have had attacks of vasoocculusive crisis with or without the knowledge of the patient, which contributed to different spectrum of surgical manifestations.

Incidence of Surgical Complications in Different Percentage of Haemoglobins in Sickle Cell Disease

Haemoglobin in gm%	No. of cases	Percentage
4-6	23	16.42%
6.1-8	44	31.42%
8.1-10	60	42.85%
10.1-12	11	7.8%
12.1+	2	1.4%

The above table shows that the majority of cases of sickle cell disease patients have moderate anaemia, i.e. 74.27% (31.42%+42.85%). Severe anaemia was detected in 23 cases (16.42%) and mild anaemia in 9.2% cases (7.8% + 1.4%)

- **Incidence of Different Surgical Complications in Sickle Cell Disease**

Surgical Complications	No. of cases	Percentage
Gall bladder and billiary tract disease	42	30.00%
Splenomegaly	72	51.42%
Hepatomegaly	56	40%
Chronic duodenal ulcer	5	3.57%
Priapism	3	2.14%
Acute abdominal crises	3	2.14%
Chronic leg ulcer	5	3.57%

Operation Performed in Sickle Cell Disease Patients during the Period of the Study

Operation	No. of cases
Ulcer debridement and skin grafting	5
Splenectomy	61
Cholecystectomy	32
Truncal vagotomy with gastro jejunostomy	5
Caverno-spongiosum shunt	3
Laparotomy	3
Total	109

One of the 140 cases, 31 cases were treated conservatively. All other cases underwent different types of operations. There were neither problems nor complications during the administration of general anaesthesia. All cases had uneventful recovery.

In this present study of **"EVALUATION OF SURGICAL COMPLICATIONS IN SICKEL CELL HEMOGLOBINOPATHY"**. 140 cases of sickle cell disease were taken into consideration.

The highest incidence of surgical complications in sickle cell disease occurred in the age group of 11 to 20 years (42.14%).The maximum number of cases were males (60%) with a male:female ratio of 8:5.Maximum number of cases presented without the sickle cell crisis (97%).Relevant family history was traced in 30% of cases.

The surgical complications of sickle cell disease like priapism chronic leg ulcer, cholelithiasis, splenomegaly, chronic duodenal ulcer, acute abdomen were found in the present series in 2.14%, 3.5%, 30%, 52.42%, 3.57%, 5% of cases respectively.

Five cases of leg ulcer were seen. Two were spontaneous and three were of traumatic origin. Skin grafting was done in 2 cases. Rest were managed conservatively with daily dressing with antiseptic lotion and antibiotic was given to control secondary infection.

Cholelithiasis was found in 42 cases and was managed surgically in 32 cases. Cholecystectomy was done. Rest were managed conservatively.

Splenomegaly was found in 72 cases and splenectomy was done in 61 cases and rest were managed conservatively. The criteria of operation for doing a splenectomy was recurrent acute spienic sequestration and chronic hyperspienism. In younger age group, the splenectomised patients were more susceptible to infections. Children who underwent splenectomy were given pneumoccal vaccine (pneumovax) and antibiotics till 18 years of age. All cases were symptom free at 2nd month of post operative period and splenectomy cases were marked with dramatic clinical improvement.

5 cases of duodenal ulceration was found. Those cases were managed surgically, truncal vagotomy with posterior gastrojejunostomy done. All were relieved of the symptoms in a follow up of 3 months.

Priaprism was seen in 3 cases and Corporaspongiosum shunt was done in these cases and the patients were relieved of their aliment.

So gall stone diseases and splenomegaly combinly accounts almost 80% of all surgical complications in sickle cell patients which were admitted to our department during the study period.

CONCLUSION

The incidence of surgical manifestations of sickle cell disease found in this study conducted by us is 0.96% , which in comparison to the earlier studies is significantly lower. This can be attributed to early diagnosis and prompt management of surgical complications and to introduction to new preventive therapies that have greatly reduced mortality and morbidity associated with surgery.

Hepatobiliary disorders and splenomegaly in SCD are common complication and about 80% of the cases were below 20 years. Abdominal pain and jaundice were among the primary presenting symptoms.

Young surgeons should be vigilant to other complications of the disease like chronic leg ulcer, priapsim and splenic complications such as acute splenic sequestration crisis. Abdominal pain in SCD represents a diagnostic challenge as its mimics many surgical emergencies and warrants careful evaluation of each patient.

It is hoped that the increasing awareness of the surgical manifestations of SCD, the introduction of minimally invasive technique and the new preventive therapies such as hydroxyl-urea aim at reducing the number and severity of sickle cell crisis by increasing synthesis of Hb-F and thus reduce the high mortality and morbidity associated with surgery in this high risk population in this part of Western Odisha where the prevalence of the disease is as high as 9%.

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