Original Article

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Splenectomy in Sickle Cell Haemoglobinopathies from a Tertiary Hospital of Southern Odisha: A Retrospective Study

Surgery Section

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ABSTRACT

Introduction: Sickle Cell Disease (SCD) is a common haemoglobinopathy, where spleen is considered to be the first organ to be involved. Although it initially undergoes enlargement, but most often there occurs autosplenectomy. But in some cases, there is persistence of splenomegaly which may be associated with various complications. In these situations, splenectomy is considered as a good option to mitigate all these morbidities.

Aim: To analyse the effect of splenectomy on haematological parameters and complications in patients of SCD in postoperative period.

Materials and Methods: This was a retrospective study conducted at SLN Medical College and Hospital, Koraput, Odisha, India, from April 2019 to March 2021. Total 42 patients of SCD underwent splenectomy. The haematological parameters {Haemoglobin (Hb), Total Leukocyte Count (TLC) and Total Platelet Count (TPC)} and blood transfusion needs were compared in preoperative and postoperative periods and analysed statistically with Student's t-test.

Results: Primary indication of splenectomy was hypersplenism 31 (73.8%) followed by splenic sequestration crisis in 8 patients (19.05%) and splenic abscess in 3 patients (7.1%). Postsplenectomy haemoglobin improvement was around 5.17 gm% (p-value<0.001). Postoperative complication was also minimal, seen in only eight patients, it was mostly due to surgical site infection (n=6) and fever (n=8). Preoperative blood transfusion rate was 9.07±1.92. Blood transfusion requirement rates were also drastically reduced to 0.5±0.707 postsplenectomy in these patients (p-value<0.001).

Conclusion: Following splenectomy, the need of blood transfusion was grossly reduced and also, the incidence of its complication was reduced.

Keywords: Hypersplenism, Sickle cell diseases, Splenomegaly

INTRODUCTION

The Sickle Cell Disease (SCD) is one of the common haemoglobinopathies characterised by an inherited structural abnormality of haemoglobin with autosomal recessive inheritance [1]. There is a mutation that leads to a single change in amino acid of haemoglobin beta chain i.e. glutamic acid is replaced with valine in 6th position [1]. Its incidence is high, where the consanguineous marriage rate is high. Mostly the first organ to be affected in SCD is spleen. Initially it is enlarged in first few years of life, but later on due to repeated microvascular occlusion it undergoes gradual progressive fibrosis leading to autosplenectomy [2]. It is frequently associated with complications like episodes of acute splenic sequestration crisis, hypersplenism, massive splenic infarction and splenic abscess [3]. The life-long prevalence of acute splenic sequestration ranges from 7% to 30% according to Topley JM et al., [4], Powell RW et al., [5], Brousse V et al., [6]. It is considered to be the second common cause of mortality in SCD following infection [4,5].

Hypersplenism is characterised by enlarged spleen, causing rapid and premature destruction of blood cells of all lineages [7]. Though, splenic abscess is rare in SCD but remains a diagnostic challenge as clinical features resembles to Vaso-occlusive crisis. Differentiating between them is important as massive splenic infarction can be managed conservatively leading to autosplenectomy [8]. Sometimes there is persistence of spleen enlargement even to adulthood. It may be accompanied by increased morbidity and mortality [8]. Elective splenectomy either open or laparoscopic is considered to be an effective measure to contain it. Study by Ghmaird A et al., revealed that following splenectomy, the blood transfusion requirement was grossly declined [9]. Mishra B et al., in his study also concluded that elective splenectomy in SCD patients can minimise the morbidity of the disease like repeated hospitalisation, blood transfusion, living with huge spleen and its accompanying symptoms and complications [10]. As the natural history of the spleen in SCD leads to autosplenectomy, in the majority of cases no surgical procedure is required.

The role of splenectomy arises whenever there is evidence of sustained hypersplenism or any life-threatening conditions like acute splenic sequestration occurs [11]. The most important question regarding splenectomy here is to what degree the surgical removal of the spleen will further increase the risk of life-threatening infection and the ideal age for operation to be performed. There is no clear answer to this question. This study was conducted with an aim to review the experience in the management of SCD with massive splenomegaly and to analyse the effect of splenectomy on haematological parameters and complication in patients of SCD in postoperative period in this tribal belt of Southern Odisha, where the prevalence of SCD is more and there is more instances of patients with persistent huge splenomegaly.

MATERIALS AND METHODS

This was a retrospective study carried out in the patients of SCD, who underwent splenectomy in the Department of General Surgery, SLN Medical College, Koraput, Odisha, India, from April 2019 to March 2021. Data was collected from hospital record section and analysed in April 2022. Approval from Institutional Ethical Committee (IEC) was obtained (03-IEC/2022 SLN MCH). It included all the patients of SCD (n=42) who were admitted for splenectomy in the department and after obtaining informed consent during the stated period of study.

Inclusion criteria: All SCD patients with persistent splenomegaly having features of hypersplenism, sequestration crisis, splenic abscess or huge splenomegaly were included in this study.

Exclusion criteria: 1) Patients of SCD not associated with above mentioned complications, 2) The very sick patients who were not fit

for surgery, 3) SCD patients with evidence of shrinking spleen size, 4) Associated with severe co-morbidity conditions were excluded from the study.

The patients were initially diagnosed and evaluated in Haematology Department, the diagnosis was on the basis of high performance liquid chromatography test and haemoglobin electrophoresis. Ultrasonogram/Computed Tomography (CT) scan of the abdomen and pelvis was done to see the spleen size, any other splenic pathology like infarction or abscess, or any incidental findings of cholelithiasis and for other abdominal pathology; if any. All the patients were vaccinated against Pneumococci, *Haemophilus influenzae* type B and Meningococci, minimum 2 to 3 weeks prior to surgery. Preoperative blood transfusion was required in all patients to bring the haemoglobin status to optimum for surgery. Open splenectomy was preferred in all of the patients, due to lack of expertise in advanced minimal invasive surgery. The immediate and early postoperative period was monitored closely for any complications. Patients were discharged on 6th-14th postoperative days. They were advised regarding vaccination schedule in postoperative period. The follow-up was done for 12 months after the surgery. Demographic data like age at surgery, sex, indication of splenectomy, preoperative and postoperative haematological parameters like Hb, TLC and TPC, frequency of blood transfusion and development of any complication or hospitalisation in the postsurgery period were noted.

STATISTICAL ANALYSIS

The preoperative and postoperative findings were compared by Student's t-tests and p-value was calculated using Statistical Package for the Social Sciences (SPSS) software version 24.0.

RESULTS

A total of 42 patients were included in this study. The mean age of subjects were 8.4±6.80 years with 71.43% of the patients presenting in the age group of <10 years [Table/Fig-1].

Age (years)	Male (n=25) n (%)	Females (n=17) n (%)	Total (n=42) n (%)
<5	7 (16.6%)	4 (9.52%)	11 (26.19%)
6-10	11 (26.19%)	8 (19.05%)	19 (45.24%)
11-15	3 (7.14%)	3 (7.14%)	6 (14.29%)
16-20	3 (7.14%)	1 (2.38%)	4 (9.52%)
>20	1 (2.38%)	1 (2.38%)	2 (4.76%)

[Table/Fig-1]: Demographic characters.

The primary indication of splenectomy was the repeated need of blood transfusion and hypersplenism in 31 patients (73.8%) [Table/Fig-2].

Indications	Number (%)
Hypersplenism	31 (73.80%)
Sequestration crisis	8 (19.05%)
Splenic abscess	3 (7.15%)

[Table/Fig-2]: Indications of splenectomy.

Splenomegaly was found in all patients with average size of 12.5±2.3 cm as measured by ultrasonography.

All the patients had undergone open splenectomy. Six patients (14.3%) had cholecystectomy together with splenectomy. Out of these three were symptomatic and rests of the patients were asymptomatic with multiple gall bladder calculi. Forty patients (95.2%) needed blood transfusion during the operation and 2 (4.8%) patients needed no blood transfusion. There was no mortality and postoperative complications were paralytic ileus in one patient and fever in eight patients (19.04%).

Average operative duration was 1.5±0.62 hours. No intraoperative mortality was recorded. Mean length of postoperative hospital stay was 7.4±3.6 days. Surgical site infection was found in six patients. Postoperatively eight patients developed fever that was managed conservatively.

In the 12 months postoperative follow-up, average haemoglobin was 10.57±0.69 gm% with a mean increase of 5.17±1.03 gm% as compared to preoperative state (p-value <0.001) for all three haematological parameters [Table/Fig-3].

Parameters	Presple-nectomy Mean±SD	Postsple-nectomy Mean±SD	Mean change±SD	p-value
Hb%	5.4±0.63	10.57±0.69	5.17±1.03	<0.001
TLC	4804.7±1292	8347± 804	3542±1488.37	<0.001
TPC	1.83±0.45	3.5±0.5	1.67±0.710	<0.001
Blood transfusion (units/year)	9.07±1.92	0.5±0.707	8.57±1.03	<0.001

[Table/Fig-3]: Changes in postsplenectomy haematological parameters at 12 month follow-up.
p-value <0.05 considered significant

Respiratory tract infection requiring hospitalisation was seen in three patients, in the 12 months follow-up period and they were managed with standard antibiotics. Painful crisis was seen in one patient on 54th postoperative day and it was managed with adequate analgesics and maintaining good hydration status. No record of Overwhelming Postsplenectomy Infection (OPSI) was noticed. When the blood transfusion was considered, preoperatively it was 9.07±1.92 blood units per year, while median rate of blood transfusion for 1st year postoperative and 2nd year postoperative was 0 and 1 respectively (p-value <0.001).

DISCUSSION

Acute Splenic Sequestration Crisis (ASSC), an important complication of SCD, results from the rapid sequestration of red blood cells in the spleen. It is the second leading cause of death after infection in the first decades of life [12]. This study revealed that eight patients had attack of ASSC (19.05%). Commonly ASSCs are seen in infants and young children, commonly between 5 months and 2 years of age [4,5,12]. However, in present study the older children were also suffered from ASSC (mean 8.4 years). Though exact cause is not known, the persistence of splenomegaly even in older age group is considered to be one of the contributing factors [13,14].

Hypersplenism is classically defined as splenomegaly with any combination of anaemia, thrombocytopenia, neutropenia (pancytopenia if all three) with compensatory bone marrow hyperplasia for a sustained period, and improvement after splenectomy [7]. Hypersplenism was the main indication for splenectomy in present study patients. Various modalities of treatment have been advocated for hypersplenism like chronic blood transfusions, partial splenectomy, percutaneous intraluminal occlusion of the splenic artery, and embolic therapy [15-17]. The main aim of these principles of treatment is to reduce the risk of OPSI, but each modality has its own limitation. However, chronic and multiple blood transfusion has risk of alloimmunisation, transmission of blood-borne infections and iron overload [18]. Again patients with iron overload disorders are susceptible to lethal infection with bacteria like *Vibrio vulnificus, Yersinia enterocolitica* and *Escherichia coli, Bacillus, Listeria* and virus like Parvo virus, Cytomegalovirus and Hepatitis B and C [19]. Three patients in present study had past history of suffering from septicaemia with pathogen like *Escherichia coli*. Apart from this repeated hospitalisation and limited availability of blood, makes it more bothersome to both parents and children.

Again, huge splenomegaly may increase the chance of splenic injury in a trivial trauma. Partial splenic embolisation may be one of the treatment options of hypersplenism, but it is unsuitable form of treatment for patients with SCD because of its associated

complications [20,21]. So, it can be said that in patients of hypersplenism, splenectomy is highly useful in decreasing their transfusion requirements and alleviating the discomfort from mechanical pressure of the enlarged spleen [14].

Splenectomy is considered to be a safe, effective and better option in the management of patients of sickle cell haemoglobinopathies with splenic complications in this region. According to the different studies, the risk of postsplenectomy sepsis is found to be increased substantially, if splenectomy is performed before 4 years of age [22,23]. However, in present study overwhelming postsplenectomy sepsis was not noticed in the 12 months postoperative follow-up period. Splenic abscess is not a common entity. Due to early autosplenectomy in patients with SCD, the chance of development of splenic abscess further reduces. In a setting of functional asplenia with repeated splenic infarction, there occurs seedling of infective agent in the persistently enlarged spleen, making them more prone to abscess formation [24]. The overall incidence of splenic abscess is 0.14-0.7% in necropsy specimen [25].

A variety of organisms can cause splenic abscess including Staphylococci, Streptococci, and gram negative bacilli like *Salmonella* and *E. coli*. Sepsis is the most common predisposing factor for splenic abscess. Three patients undergone splenectomy for splenic abscess in present study and all were adults. They presented with fever and abdominal tenderness. Multilocular splenic abscess were found in the above patients of present study group and the size ranged from 3 cm to 12.5 cm. It is recommended that in unilocular abscesses of <3 cm in size, drainage under radiological guidance with proper antibiotic coverage is a good and safe option [24]. When abscess size is >10 cm or non surgical approach is not helpful, splenectomy has been considered to be next method of treatment [13,26].

In present study, the mean length of hospital stay after the operation was 7.4±3.6 days, compared to other study where it was three days for laparoscopic splenectomy [27]. When the haematological parameters were compared it was found that, the mean preoperative Hb was 5.4±0.63 gm/dL and postoperatively it was increased to 10.57±0.69 gm% (p-value <0.001). According to Lesher AP et al., there was 38% decrease in the number of units transfused during first six months postsplenectomy and a 45% decrease during 6 to 12 months postsplenectomy and all the haematologic parameters remained stable or improved [22]. Red blood cell turnover was decreased as evidenced by raised haematocrit and decreased reticulocytes count after the splenectomy. All these above factors indicated that the survival of the transfused red cells were greatly improved [22]. Another Saudi Arbian study by Al Salem AH clearly showed that the postoperative haematocrit and reticulocytes significantly improved in children following splenectomy for hypersplenism [13]. Svarch E et al., also noticed that haemoglobin concentration was significantly improved (6.0 vs. 7.7; p-value=0.01) along with reduction in blood transfusion after partial splenectomy for acute splenic sequestration [28]. In present study, there was history of one unit transfusion in one patient only which clearly demonstrated the advantage of splenectomy in SCD patients.

There are several advantages of splenectomy in SCD apart from improvement in haematological variables. Due to improved red cell half life, haematopeotic pressure over bone marrow is greatly reduced [29]. There is relief of abdominal heaviness from enlarged spleen. Patient develops a good appetite as gastric compression by spleen is no more present. They are not transfusion dependent. All these factors help in regaining, a well appreciated growth in postsplenectomy patients [30]. With availability of good surgical technique and safe anaesthetic option and with above mentioned advantages following splenectomy, it can be opined that elective splenectomy in SCD with persistent enlarged spleen or having sequestration crisis is very much safe and beneficial to these patients. It is also supported by various studies [13,22,29]. However, this study was conducted in an area where rate of consanguineous marriage is very high and the incidence of persistence of splenomegaly in SCD is also more. Due to poor standard of living, conservative approach to spleen could not be considered as a better option to them.

Limitation(s)

The limiting factor in this study was the small study population, and study involving only a limited geographical area. It will be better to study a large numbers of patients from a wider geographical area, with variable presentation to reach an acceptable inference.

CONCLUSION(S)

With good perioperative care, well planned vaccination schedule and appropriate antibiotic prophylaxis was required. An elective splenectomy in SCD is not only considered to be safe but also reduces the morbidity and life-threatening complications associated with it. It also significantly reduces the need of repeated blood transfusion and its related disadvantages. The incidence of OPSI is also very minimal with adequate vaccination coverage.

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