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Splenectomy and Major Sickle Cell Syndromes in Ouagadougou: What is the Benefit for Patients after Surgery?

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Abstract

Introduction: Splenectomy in major sickle cell syndromes have specific indications. The main one is hypersplenism. The aim of our research was to study and to compare the clinical and hematological parameters pre and postoperatively.

Materials and Methods: This was a retrospective cohort study with an analytical aim of splenectomy performed in patients with major sickle cell syndromes from 2010 to 2019 at two hospitals (University hospital center of Yalgado Ouédraogo and Saint Camille Hospital of Ouagadougou) in Ouagadougou.

Results: In 10 years, 31 cases were collected. The mean age of the patients was 27.6 ± 2 years. A history of at least three vaso-occlusive crises per year was noted in all cases. Before splenectomy, clinical anemia was noted in all cases. Physical examination revealed painful splenomegaly in 19 cases (61.3%). Hemoglobin phenotypes were: 16 cases (SC), 10cases (SS), 5 cases ($S\beta^+$). The average blood transfusion was 6 ± 2/person/year. Splenectomy was indicated for repeated attacks of hypersplenism in 18 cases. Twenty-nine patients underwent total splenectomy and two underwent partial splenectomy. In 40.8% of cases, splenectomy was associated with cholecystectomy. In one month postoperatively, the hematological parameters showed clear improvement with an increase in the hemoglobin level of 3.7 points (p=0.001), red blood cells (p=0.03) and platelets (p=0.02). Morbidity was 3.7% and mortality 0%.

Conclusion: Splenectomy decreases the postoperative transfusion needs in the short and medium term. A good follow-up limits postoperative complications.

Keywords: Major-sickle-cell-syndrome; Splenectomy; Hypersplenism; Ouagadougou

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Introduction

Described for the first time by Herrick in 1910, sickle cell disease is a hereditary hemoglobin disease that is widespread throughout the world, particularly in Africa and Asia. It is due to the mutation of the sixth codon of the β-globin chain, at the origin of the synthesis of Hemoglobin S (HbS). The abnormality in hemoglobin synthesis can be qualitative or quantitative [1]. Homozygous sickle cell disease SS and composite heterozygosity SC, S-β-thalassemia, SOArab, SDPunjab, are grouped under the term major sickle cell syndromes [2]. Nowadays, early detection and greater understanding of the pathophysiology of these syndromes have led to improved management [2]. Indeed, the life expectancy of patients has improved according to several authors [1-3]. Consequently, the appearance of chronic pathologies in the cardiovascular, osteoarticular, ophthalmic and digestive systems is increasingly described in the literature [1,3]. The spleen is one of the organs affected by the progressive digestive complications of this disease [2]. This damage is manifested by hypersplenism, which is reflected by the progressive increase in the volume of the spleen during the first decades [4,5]. Persistence of hypersplenism in old age would be the cause of

various complications such as sequestration, infarction and splenic abscesses, which increase morbidity and mortality (20%) in these subjects, hence the indication for splenectomy [3,5,6]. However, this indication remains controversial because of the essential role of the spleen in the immune system and the risks of sepsis incurred by its removal [1,2,5]. The aim of our research was to study the impact of splenectomy on the life of the patients by comparing the clinical and hematological parameters pre- and postoperatively in our context.

Materials and Methods

This was a retrospective cohort study, with an analytical aim, on splenectomies performed in patients with major sickle cell syndromes, from 2010 to 2019 at the Yalgado Ouédraogo University Hospital and the Saint Camille Hospital in Ouagadougou. Our study included all the records of patients with sickle cell disease received in surgical consultation in the two visceral surgery departments for whom the indication for splenectomy was given during the study period. Not included in our study were all the files that did not contain sufficient information on the parameters studied and those of patients lost to follow-up at less than one year. For each patient, the parameters studied were:

- **Socio-demographic data:** Age, sex, occupation, origin
- **Clinical data:** Weight, height, Body Mass Index (BMI), number of vaso-occlusive crises lasting more than 72 h, Hackett classification of splenomegaly, presence or absence of associated visceral complications.
- **Paraclinical data:** Pre- and postoperative blood count, hemoglobin phenotypes and results of ultrasound and/or abdominal CT scan.
- **Therapeutic and evolutionary data:** Type of treatment, number of blood transfusions per person per year before and after splenectomy; indications and type of splenectomy, spleen weight, duration of hospitalization and follow-up, occurrence of complications, mortality, type of vaccine received, malaria prevention. The indication for splenectomy was established by a multidisciplinary team including hematologists, surgeons and anesthesiologists in view of the high number of hypersplenism attacks, transfusion requirement, and other complications.
- All patients were prepared for the procedure in the general surgery departments of both hospitals and were followed for at least one year after surgery. They also had regular appointments with their hematologists. They all received 4 types of vaccine (pneumo 23, Menevax (ACYW135), TyphimVi and hepatitis B) at least one month before surgery. Antibiotic prophylaxis and malaria prevention were also performed (use of insecticide-treated nets and sulfadoxine-pyrimethamine-based chemoprophylaxis). Hydroxyurea was prescribed to SS homozygous patients.
- The assessment of the postoperative evolution was possible thanks to the comparison of clinical, biological and therapeutic data.
- Data were collected using a collection form, then entered and processed using Word and Excel software, and exported to Stata software version 14.0 for statistical analysis. We considered the data from the analysis to be significant at the 5% level. The paired t-test allowed us to compare the averages of the blood count parameters in pre- and post-operation and the frequencies of blood transfusion before and after splenectomy were compared using the McNemar χ^2 test.

Results

Epidemiological data

Over a period of10 years, 31 patients with major hemoglobinopathies underwent splenectomy in the visceral surgery departments of both hospitals. The mean age of the patients was 27.6 ± 2 years, with extremes of 15 and 51 years; 18 of whom were male and 13 female (sex ratio 0.7). Twenty-two (22) patients were older than 20 years.

Clinical data

Seventeen patients (54.8% of cases) were classified as WHO performance status stage 2 and 12 patients (38.7% of cases) as stage 3. The mean body mass index (BMI) was 19.8 kg/m^2 with extremes of 18 and 20.8 Kg/m^2. Delayed growth and development was noted in 32.3% of cases (10 cases). A history of at least three vaso-occlusive crises lasting more than 72 h associated with an acute chest syndrome was noted in all cases. Clinical anemia was noted in all cases prior to surgery. It was characterized by conjunctivo-palmoplantar pallor and respiratory difficulties. Conjunctival jaundice was noted in 74.1% of cases. Physical examination revealed painful splenomegaly in 19 cases (70.4%). Hackett's classification was used for splenomegaly. Table 1 summarizes the distribution of patients according to this classification. In 48.4% of cases, splenomegaly was associated with hepatomegaly and in 45.2% of cases with a palpable gallbladder.

Paraclinical data

The mean baseline hemoglobin level was 10.1 ± 1 g/dl. In the immediate preoperative period, the mean hemoglobin level was 6.1 ± 1.8 g/dl. Decreased red blood cells were noted in all cases, with a mean of 3.198.106/ml ± 686.475. Eleven patients had leukopenia (35.5% of cases). Thrombocytopenia with platelets less than 100.000/mm^3 was noted in 7 patients (22.6% of cases). Hemoglobin phenotypes consisted of 16 cases (51.6%) of composite heterozygote's SC, 10 cases (32.3%) of homozygote SS, and 5 cases (16.1%) of Sβ+ sickle cell disease. Before splenectomy, the average frequency of blood transfusion per year and per person was 6 ± 2.5. The distribution of patients according to the frequency of blood transfusion per year and per person was listed in Table 2. Abdominal-pelvic ultrasonography and abdominal-pelvic CT scan showed splenomegaly in 100% of cases, hepatomegaly in 48.4% of cases, and a large gallbladder with lithiasis content in 45.2% of cases. Splenic infarction (Figure 1a), splenic

Table 1: Distribution of patients according to frequency of blood transfusions before splenectomy.

Number of blood transfusions	Number of cases	Percentage (%)
<3	2	6.5
3-6	11	35.4
>6	18	58.1
Total	31	100

More than 50% of patients needed more than 6 blood transfusions per year

Table 2: Distribution of patients according to the Hackett classification of splenomegaly.

Hackett's classification	Number of cases	Percentage (%)
Splenomegaly type 3	7	22.6
Splenomegaly type 4	15	48.4
Splenomegaly type 5	09	29.0
Total	31	100

Hackett's type IV splenomegaly was the most common

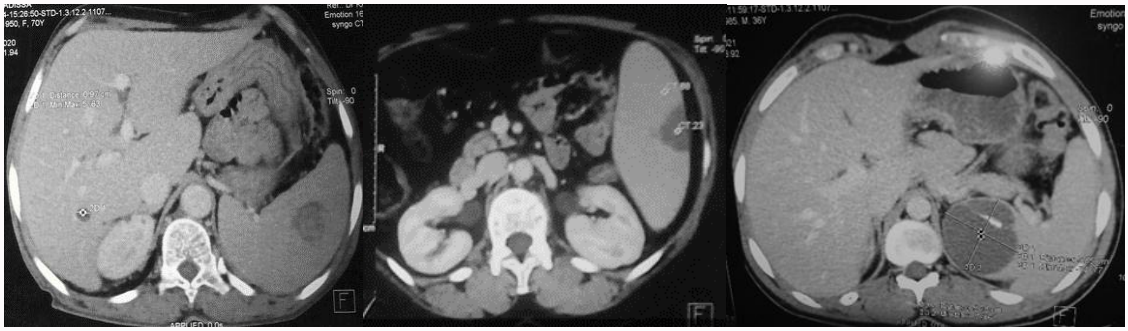

Figure 1: a) Splenic infarction. b) Splenic abscess. c) Splenic cyst.

Table 3: Comparison of preoperative, one month and one year postoperative hematological data.

Hematological data	Variations					
	Preoperative-M1 postoperative		Preoperative-A1 postoperative		M1-A1postoperative	
	Mean Diff	P	Mean Diff	P	Mean Diff	P
Hb (g/dl)	+3.7	0.001	+2.3	0.02	-0.74	0.002
GR (10^6/ml)	+1.8	0.032	+1.1	0.045	-0.67	0.02
GB (10^3/ml)	+0.2	0.056	+0.8	0.27	-0.43	0.16
Plaq (10^3/ml)	+46.8	0.02	+44.5	0.008	-1.83	0.9

Mean Diff: Difference in mean; M1: One month in post operative; A1: One year in post operative

Table 4: Changes in the frequency of pre- and post-splenectomy transfusion requirements.

Blood transfusion Annual frequency	Before splenectomy		After splenectomy (one year)		
	Number of cases	Percentage %	Number of cases	Percentage %	P
<3	2	6.5	22	71	0.01
3-6	11	35.4	8	25.8	0.58
>6	18	58.1	1	3.2	0.06
Total	31	100	31	100	

abscess (Figure 1b) and splenic cyst (Figure 1c) were diagnosed in 4; 2 and 1 case respectively.

Therapeutic data

Splenectomy was indicated for repeated attacks of hypersplenism (18 cases), splenic sequestration (6 cases), splenic infarction (4 cases), splenic abscess (2 cases) and splenic cyst (1 case). The approach was a median above and below the umbilical in 20 cases and a left subcostal in 11 cases. No laparoscopic splenectomy was performed. Twenty-nine patients (80.6%) had total splenectomy (10 SS, 15 SC, and 4 Sβ+ cases) and two patients had partial splenectomy (1 Sβ+ and 1 SC case). In 40.8% of cases, splenectomy was associated with cholecystectomy for vesicular lithiasis. The mean spleen weight was 790 g with extremes of 400 g and 1970 g. The Anatomopathology of the surgical parts did not show any malignancy.

Evolutionary data

The immediate postoperative evolution was simple without notable complications in all cases. In one month postoperatively, the hematological parameters showed a clear improvement with an increase in the hemoglobin level of 3.7 points (p=0.001), red blood cells (p=0.03) and platelets (p=0.02). The one-year postoperative check-up showed a decrease in the hemoglobin level, which nevertheless remained higher than the baseline hemoglobin level. The comparison of the mean hematological parameters preoperatively, at one month and at one year postoperatively is shown in Table 3. The number of blood transfusions decreased significantly postoperatively. The comparison of the evolution of the frequency of blood transfusions before and after splenectomy is represented by Table 4. The average hospital stay was 6 ± 3.2 days. Post-splenectomy morbidity was marked by common germ bronchopneumonia noted in two patients, three weeks and one month after splenectomy. It was successfully treated as an outpatient with amoxicillin + clavulanic acid for 15 days. Recurrence of chronic anemia was also observed in 7 patients. Mortality was nil in our series with a mean follow-up of 3.6 years.

Discussion

Over a period often years, 31 patients with major sickle cell syndromes underwent splenectomy in our two visceral surgery departments, an annual frequency of 3 cases per year. The mean age of the patients was 27.6 ± 2 years. All these patients presented at least three vaso-occlusive crises associated with an acute chest syndrome. All of them had splenomegaly classified as Hackett stage IV (48.4%) on clinical examination. Eighteen patients (58.1%) had received more than six transfusions per year. In the literature, splenic damage is one of the visceral complications observed in patients with major sickle cell syndromes [1,2,6]. In the majority of cases, it is manifested by hypersplenism and splenic sequestration attacks, which have been described as the main indications for splenectomy in these patients [6-8]. However, these indications have been the subject of several controversies in the literature, due to the important role of the spleen in the immune system [1,3,6]. Indeed, Owusu [1] in his meta-analysis noted that splenectomy should be contraindicated especially

during the first years of life. Ahmed made the same observation in their review of the literature [9]. In our series, all patients were adolescents and adults. The main indication for splenectomy was hypersplenism (58.06% of cases), followed by splenic sequestration crises (19.4% of cases). Dick et al. [10] and Ghmaird et al. [11], in their series of 11 and 24 cases respectively, noted that the main indication for splenectomy was hypersplenism. However, in the series of Koffi (21 cases), increased transfusion requirements were the main indication for splenectomy [11]. However, it was noted that there was a concordance between our different results, as the first manifestation of hypersplenism would be increased transfusion requirements [1-3]. In our series, 29 cases of total splenectomy were reported against 2 cases of partial splenectomy. The fear of recurrence of hypersplenism and splenic sequestration attacks would have motivated the choice of this radical treatment which was made in a multidisciplinary consultation meeting. In addition, there are problems related to blood transfusion such as blood shortages, the risk of all immunization and contamination, especially by hepatitis B and C encountered in our context. Other authors have made the same observation [3,9]. This therapeutic attitude has been adopted by several practitioners [3,10,12]. The two cases of partial splenectomy involved Sβ+ thalasso-sickle cell subjects. Indeed, conservative surgery has been recommended with a view to preserving splenic function as long as possible [1,6,11]. The mean spleen weight was 790 g with extremes of 400 g and 1970 g. According to El-Salem et al. [3], large spleens (1000 g on average) alone would constitute an indication for splenectomy. In 40.8% of cases splenectomy was associated with cholecystectomy for vesicular lithiasis. In their series, Ghmaird et al. [11] had noted an association of these procedures in 29.2% of cases. Postoperative morbidity in the short and medium term represented 6.5% of the cases in our study. It was a bronchopneumopathy diagnosed and successfully treated in two cases in ambulatory care. Other authors have noted higher rates of postoperative complications [3,11,12]. The average hospital stay was 6 ± 2 days. The laparoscopic approach, if used in our series, could have shortened this stay. In fact, according to several authors, laparoscopic splenectomy would confer a significant benefit to the patients, hence shortening the hospital stay to 3 days on average [4,9,11]. In one month postoperatively, the hematological parameters showed clear improvement with an increase in hemoglobin level of 3.7 points (p=0.001), an increase in red blood cells (p=0.03) and platelets (p=0.02). Our results are comparable to those in the literature. In addition, Ghmaird [10] and Koffi [11] noted in their series a significant increase in hemoglobin and platelet levels in the short and medium term. In our series, the hematological check-up at one year post-op showed a decrease in the hemoglobin level, which nevertheless remained higher than the baseline hemoglobin level (p=0.02). The observation of a long-term decrease in hemoglobin level after splenectomy has been commonly reported in the literature [9,11,12]. Some authors [5,11-13] have noted the recurrence of chronic anemia in certain patients, as was the case in our series. Indeed, in our series, the comparative study of the frequency of blood transfusion pre and post operatively showed a significant decrease post operatively (p=0.01). All our patients received 4 types of vaccine (pneumo 23, Menevax (ACYW135), TyphimVi and hepatitis B) at least one month before surgery. All patients also received malaria prevention (use of insecticide-treated nets and sulfadoxine-pyrimetamine chemoprophylaxis). This may partly account for the low percentage of postoperative complications that we found. Hydroxyurea was prescribed to our SS homozygous patients not only to help maintain a high fetal hemoglobin level, but also to prevent the acute pain syndrome. In the literature, some authors have adopted this therapeutic attitude [13]. All our patients were regularly followed for at least one year in the surgical department. There were no deaths in our study with a 2-year follow-up.

Conclusion

The indication of splenectomy has not been a common practice in major sickle cell syndromes in our context. The decision was taken in a multidisciplinary consultation meeting with the aim of palliating the serious complications related to acute sequestration attacks and hypersplenism. Splenectomy improved hematological parameters in the medium term in all cases. Vaccination therapy and antimalarial chemoprophylaxis should be performed routinely in our setting to prevent the occurrence of possible postoperative complications.

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