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Assessment of Spleen by Ultrasound in Sickle Cell Patient among Indian Population

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ABSTRACT

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Background- Ultrasonography is one of the non invasive, cheap, readily available and reliable method for assessment of not only spleen size, but also the echotexture of spleen in cases with sickle cell disease. This study was conducted to sonographically assess and compare the spleen in patients with sickle cell anemia and age and sex matched controls among the Indian population.

Methodology- The study was conducted as an observational comparative study on a total of 80 confirmed cases of Sickle cell anemia and 80 controls at tertiary care centre. Detailed history regarding sociodemographic variables such as age and sex was obtained from all the cases and controls. All the study participants were subjected to ultrasound examination and length, width and echotexture was noted.

Results- A total of 80 cases with sickle cell anemia and matched controls with mean age of 10.59±1.18 years. Mean length as well as width of spleen was found to be significantly higher in cases as compared to controls ($p<0.05$). Also we noted that all the controls had normal splenic echotexture whereas splenic echotexture was altered in majority of cases with sickle cell disease as described in table 2 ($p<0.05$).

Conclusion- Splenic abnormalities and complications are common in patients with SCD. While enlarged spleen size is seen in young cases, autosplenectomy, shrunken spleen, infarction, abscess and other changes in echotexture are commonly observed in patients with advancing age. Sonological assessment might help in assessing the splenic complication in patients with SCD and thus must be routinely done in such cases.

KEYWORDS:

sickle cell anemia, autosplenectomy, spleen size, ultrasonography, spleen echotexture.

INTRODUCTION

Sickle cell disease is an autosomal recessive disorder, which is characterized byalteration in normal globin chain of hemoglobin, and formation of abnormal hemoglobin chain (HbS), due to point mutation at the sixth position of the beta-globin chain, where glutamine is replaced by valine.This result in rigid, abnormal and sickle shaped red blood cells (RBCs).[1]Approximately 90% of the total hemoglobin is HbS in cases with sickle cell anemia and this level has direct association with the severity of symptoms.[2]In this form of hemoglobinopathy, the patients present with anemia, sickling crisis in the form of organ ischemia, acute painful episodes and chronic organ damage, which has significant impact on life expectancy.[3] Literature suggest that the sickle cell disease is more prevalent in Sub Saharan Africa.[4] In India, the condition is commonly observed in tribal populations of Central India and northern Kerala and Tamil Nadu, also called the 'sickle cell belt'.[2] Sickle cell disease in Indian population is found in 1 per 16000 screened cases.[5]

Splenic manifestations are commonly reported in patients with sickle cell disease. It has been estimated that approximately two third of patient with SCD present with splenic manifestations by the 2 years of age.[6]These splenic changes may be structural

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or functional, ranging from non functional splenomegaly to splenic infarction and splenic abscess.Among various splenic manifestations, splenomegaly and autosplenectomyare common and observed later in life.Splenic manifestations in SCD are attributed to decreased levels of hemoglobin F (HbF), high level of irreversible sickle cells (ISC), chronic malarial infection and increased level of antibody (IgG and IgM).[7]As a result of these splenic changes, there occurs hypersplenism, acute splenic sequestration, and higher susceptibility to the infection.[8]

Routinely, per-abdomen examination is done to assess the splenomegaly, but this method is not reliable. Ultrasonography is one of the non invasive, cheap, readily available and reliable method for assessment of not only spleen size, but also the echotexture of spleen in cases with sickle cell disease. The present study was conducted to sonographically assess and compare the spleen in patients with sickle cell anemia and age and sex matched controls among the Indian population.

MATERIALS AND METHODS

The study was conducted as an observational comparative study on a total of 80 confirmed cases of Sickle cell anemia and 80 age and sex matched controls were recruited at Department of Radiodiagnosis NSCB Medical College Jabalpur. Patients with sickle cell anemia were recruited from outpatient sickle cell clinic present at our study area whereas the age and sex matched patients were recruited from other outpatient Departments of our study centre.

Inclusion criteria

- Cases
    - Patients of any age and sex with sickle cell disease, attending sickle cell clinic.
    - Patients/ relatives giving consent for the study
- Controls
    - Patients with diseases other than sickle cell disease
    - Willing to participate in the study and undergo ultrasonography

Exclusion criteria

- Patients with other conditions affecting the spleen such as malaria, lymphomas, leukemias, myeloproliferative disorder, thalassemia, other hemolytic anemias.
- Participants with history of previous splenectomy
- Not willing to participate in the study

Both verbal and written informed consent was obtained from the study participants belonging to more than 18 years of age whereas the consent was taken from parents or guardians of the minor participants.

After obtaining ethical clearance from Institute's ethical committee, all the participants (cases and controls) fulfilling the inclusion criteria were included in our study. Detailed history regarding sociodemographic variables such as age and sex was obtained from all the cases and controls. All the study participants were subjected to ultrasound examination using B-mode real-time transabdominal ultrasound scan. A3.5–5 MHz curvilinear transducerof AlpinionE-CUBE-i7 is used All the cases and controls were examined sonographically in supine position and the length as well as width of spleen was measured by the single assessor. The length of spleen was measured at the level of hilum in mid clavicular line. When the spleen was not visualized at the position of spleen, it was defined as autosplenectomy,[9-11] All the measurements were taken on the image obtained in a static freeze screen using the electronic caliber. Apart from this, the echotexture of spleen was also assessed and findings were documented.

Statistical analysis-All the data was obtained in the proforma and entered in MsExcel. Analysis of data was done with the help of IBM SPSS software version 20 for Windows (IBM, Chicago, Illinois, USA). The width and length of spleen was expressed in mean and standard deviation and was compared between cases and controls using independent t test whereas echotexture of spleen was expressed as frequency and proportion and compared between cases and controls using chi square test. P value of less than 0.05 was considered as statistically significant.

RESULTS

A total of 80 cases with sickle cell anemia and matched controls were included in our study. Mean age of cases and controls was 10.59±1.18 years.

Table 1- Comparison of cases and controls according to age and sex

Sociodemographic variables		Cases (n=80)	Controls (n=80)	P value
Age (years)	≤10	16 (20%)	16 (20%)	1.0
	11-20	29 (36.2%)	29 (36.2%)	
	21-30	23 (28.7%)	23 (28.7%)	
	31-40	8 (10%)	8 (10%)	
	>40	4 (5%)	4 (5%)	

Sex	Male	37 (46.2%)	37 (46.2%)	1.0
	Female	43 (53.8%)	43 (53.8%)	

Majority of cases belonged to 11 to 20 years of age (36.2%), followed by 28.7% cases belonging to 21 to 30 years of age. About 46.2% cases were males. We found no significant difference in age and sex composition between cases and controls and thus, cases and controls were comparable with respect to age and sex (p>0.05).

Table 2- Comparison of sonographic spleen parameters between cases and controls

Spleen on sonography		Cases (n=80)	Controls (n=80)	P value
Length (mm)	Mean±SD	106.85±46.43	83.16±13.19	**0.001**
Width (mm)	Mean±SD	48.51±24.0	35.5±5.68	**0.001**
Spleen echotexture	Normal	2 (2.5%)	80 (100%)	**0.001**
	Coarse & hyperechoic	19 (23.8%)	0 (0%)	
	Fibrotic and littered	17 (21.2%)	0 (0%)	
	Infarct and abscess	8 (10%)	0 (0%)	
	Fine and hyperechoic	11 (13.8%)	0 (0%)	
	Infarct/ abscess/ fibrotic and littered	3 (3.8%)	0 (0%)	
	Shrunken calcified	5(6.25%)	0 (0%)	
	Autosplenectomy	15 (18.8%)	0 (0%)	

Autosplenectomy was found in 15 cases with sickle cell anemia and hence length and width could be assessed in 65 cases. Mean length as well as width of spleen was found to be significantly higher in cases as compared to controls (p<0.05). Also we noted that all the controls had normal splenic echotexture whereas splenic echotexture was altered in majority of cases with sickle cell disease as described in table 2 (p<0.05).

Table 3- Age-wise comparison of sonographic spleen parameters between cases and controls

Spleen on sonography		Age (years)	Cases	Controls	P value
Length (mm)	Mean±SD	≤10	80.29±19.5	67.88±9.4	**0.031**
		11-20	36.71±6.7	30.88±4.2	**0.008**
		21-30	114.70±47.9	81.97±7.1	**0.001**
		31-40	52.39±23.1	35.17±3.5	**0.001**
		>40	104.85±47.1	87.87±9.4	**0.042**
Width (mm)	Mean±SD	≤10	50.55±26.9	36.87±4.4	**0.021**
		11-20	141.0±66.1	101.0±12.9	0.116
		21-30	61.8±33.4	38.5±8.8	0.080
		31-40	120.0±0	90.25±17.0	0.080
		>40	26.0±28.3	43.25±9.54	0.291
Echotexture	Normal	≤10	2 (12.5%)	16 (100%)	**0.001**
	Coarse & hyperechoic		3 (18.8%)	0 (0%)	
	Fibrotic and littered		2 (12.5%)	0 (0%)	
	Fine and hyperechoic		7 (43.8%)	0 (0%)	
	Autosplenectomy		2 (12.5%)	0 (0%)	
	Normal	11-20	0 (0%)	29 (100%)	**0.001**
	Coarse & hyperechoic		6 (20.7%)	0 (0%)	
	Fibrotic and littered		6 (20.7%)	0 (0%)	
	Infarct and abscess		4 (13.8%)	0 (0%)	
	Fine and hyperechoic		4 (13.8%)	0 (0%)	
	Infarct/ abscess/ fibrotic and littered		1 (3.4%)	0 (0%)	

	Shrunken calcified		2 (6.9%)	0 (0%)	
	Autosplenectomy		6 (20.7%)	0 (0%)	
	Normal	21-30	0 (0%)	23 (100%)	**0.001**
	Coarse & hyperechoic		7 (30.4%)	0 (0%)	
	Fibrotic and littered		5 (21.7%)	0 (0%)	
	Infarct and abscess		4 (17.4%)	0 (0%)	
	Infarct/ abscess/ fibrotic and littered		1 (4.3%)	0 (0%)	
	Shrunken calcified		3 (13%)	0 (0%)	
	Autosplenectomy		3 (13%)	0 (0%)	
	Normal	31-40	0 (0%)	8 (100%)	**0.001**
	Coarse & hyperechoic		2 (25%)	0 (0%)	
	Fibrotic and littered		3 (37.5%)	0 (0%)	
	Infarct/ abscess/ fibrotic and littered		1 (12.5%)	0 (0%)	
	Autosplenectomy		2 (25%)	0 (0%)	
	Normal	>40	0 (0%)	4 (100%)	**0.046**
	Coarse & hyperechoic		1 (25%)	0 (0%)	
	Fibrotic and littered		1 (25%)	0 (0%)	
	Autosplenectomy		2 (50%)	0 (0%)	

As observed from the above table, there was significant difference in mean length of patients stratified based on age across all age range ($p<0.05$). We observed significant difference in width of spleen between cases and controls in less than 10 years of age ($p<0.05$). However, echotexture was affected in significantly higher proportions of cases as compared to controls across all age range ($p<0.05$).

Table 4- Sex-wise comparison of sonographic spleen parameters between cases and controls

Spleen on sonography		Sex	Cases	Controls	P value
Length (mm)	Mean±SD	Male	105.15±43.1	83.09±13.4	**0.002**
		Female	47.94±23.2	35.23±5.9	**0.001**
Width (mm)	Mean±SD	Male	108.59±50.3	83.24±13.1	**0.004**
		Female	49.09±25.2	35.89±5.5	**0.003**
Echotexture	Normal	Male	1 (2.7%)	37 (100%)	**0.001**
	Coarse & hyperechoic		9 (24.3%)	0 (0%)	
	Fibrotic and littered		1 (2.7%)	0 (0%)	
	Infarct and abscess		9 (24.3%)	0 (0%)	
	Fine and hyperechoic		5 (13.5%)	0 (0%)	
	Infarct/ abscess/ fibrotic and littered		5 (13.5%)	0 (0%)	
	Shrunken calcified		2 (5.4%)	0 (0%)	
	Autosplenectomy		5 (13.5%)	0 (0%)	
	Normal	Female	1 (2.3%)	43 (100%)	**0.001**
	Coarse & hyperechoic		10 (23.3%)	0 (0%)	
	Fibrotic and littered		7 (16.3%)	0 (0%)	
	Infarct and abscess		3 (7%)	0 (0%)	
	Fine and hyperechoic		6 (14%)	0 (0%)	
	Infarct/ abscess/ fibrotic and littered		3 (7%)	0 (0%)	
	Autosplenectomy		10 (23.3%)	0 (0%)	

As observed from the above table, there was significant difference in mean length, width as well as echotexture between cases and controls when we stratified patients based upon gender ($p<0.05$).

DISCUSSIONS

Sickle cell disease is a genetic disorder characterized by alteration in normal globin chain, resulting in production of abnormal hemoglobin chains. The altered structure of hemoglobin or sickling of cell lead to vascular occlusion and thereby cause ischemia in the multiple organs.[12] Spleen is commonly affected in patients with sickle cell anemia and is one of the early organs to be affected in this condition. The spleen is found to be enlarged initially especially during the first decade of life, but due to repeated episodes of vasoocclusion and infarction as a result of crisis, there occurs progressive atrophy of spleen, ultimately leading to autosplenectomy. However, in some cases, splenomegaly persist till older age group and present with certain complications such as hypersplenism, sequestration crisis, splenic infarction and abscess, necessitating the need for splenectomy.[13-15] Splenic complications in cases with sickle cell anemia are significantly associated with increased morbidity as well as mortality. Thus, it is essential to assess splenic changes in these patients.[16]

Ultrasonography is an easily available, simple, affordable, and non invasive imaging modality that plays an important role in detection of splenic changes early.[17]In present study, a total of 80 cases with SCD were included, and mean age of patients was10.59±1.18 years (Range 2 to 52 years), majority belonging to 11 to 20 years of age (36.2%). We reported slight female predominance for sickle cell anemia with male:female ratio of 0.86:1. Cases and controls were comparable with respect to age and sex ($p>0.05$). However, in a study of Luntsi et al, majority of cases with sickle cell disease belonged to 10 to 16 years of age (56.8%) and highest preponderance of complications associated with sickle cell disease was present in patients belonging to 17 to 23 years of age. Also they reported higher incidence of sickle cell disease was reported in females (57.9%).[17]On the other hand, in a study of Eze et al, majority (19.42%) cases belonged to 7 to 11 years of age and about 53.4% cases were females.[10]Similarly in a study of Ugwu et al, the authors included 237 cases with SCA with equal number of controls and majority of cases belonged to 1 to 10 years of age (53.6%); with 53.6% males.[18]

Ultrasonographically, we assessed spleen size (length and width) and echotexture of spleen. We found mean length as well as width of spleen to be significantly higher in case with SCA as compared to controls ($p<0.05$). The length of spleen was found to be significantly higher in patients with sickle cell disease in all age range ($p<0.05$),and both the genders whereas width of spleen was significantly higher in children with SCA as compared to controls below 10 years of age ($p<0.05$). Mean width was found to be significantly higher in cases in both males as well as females ($p<0.05$). Also, the echotexture was altered in significantly higher proportions of cases as compared to controls ($p<0.05$), irrespective of age and sex of the patients.

Our study findings were supported by the findings of Eze et al, in which the authors reported significantly higher mean length and width in cases with SCD as compared to controls (97.67 ± 39.61 mm vs. 80.84 ± 16.89 mm; and 47.28 ± 20.68 vs. 39.38 ± 9.39 mm respectively; $p<0.05$).[10]In this study by Eze et al, the authors reported significant variation in spleen size in SCD patients during 2 years to 18 years of age, beyond which, the patients with reduced and increased spleen size, both were seen, and maximum cases with shrunken spleen/ auto-splenectomy were observed in 18 to 33 years of age.[10]

Our study findings were also concordant to the findings of Luntsi et al, in which the authors reported splenomegaly (24.9%), auto splenectomy (17.9%), increased echotexture (7.25%) and calcified spleen (8.7%) in significantly higher proportions of cases with SCD as compared to controls ($p<0.05$).[17] Similarly, Ugwu et al reported significantly higher mean length of spleen in cases with SCA as compared to cases in patients across all age range when stratified based upon age, except below 10 years of age.[18]Splenomegaly in cases with SCD is attributed to sequestration syndrome, which is a common splenic complication, consisting of rapid pooling of blood in the spleen and thus resulting in depletion of intravascular volume.[19]

Our study had certain limitations. First, the age and sex matched controls were included in our study, however other confounders may have been missed. The controls were selected from hospital, which may be associated with bias. The sample size of the study was small, only 80 cases and 80 controls of varying age range were enrolled and the study was unicentric.

CONCLUSION

Assessment of spleen routinely with the help of sonography in patients with sickle cell disease may help in management of these patients. Splenic abnormalities and complications are common in patients with SCD. While enlarged spleen size is seen in young cases, autosplenectomy, shrunken spleen, infarction, abscess and other changes in echotexture are commonly observed in patients with advancing age. Sonological assessment might help in assessing the splenic complication in patients with SCD and thus must be routinely done in such cases.

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