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Original Research Article

Splenomegaly in children

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ABSTRACT

Background: Splenomegaly occurs when the size of the spleen is increased by cells or tissue components or by vascular engorgement. In childhood, it is generally first suspected upon physical examination. The aim of the present study was to find out the prevalence and possible cause of splenomegaly in children admitted in pediatric ward and NICU at tertiary care center.
Methods: In this study, total 124 children of age between 0-12 years with clinically palpable splenomegaly, admitted to the wards were studied during the period of 18 months. A detailed history, thorough clinical and all relevant investigation was done. The enlargement of the spleen was graded as per Hacketts and conventional classification. The prevalence, cause of splenomegaly and outcome of the study was noted.
Results: The prevalence of splenomegaly was 1.46%. Most common grade of splenomegaly was grade III (33%) of Hackett's classification. The most common presenting symptom was fever (75%) and sign was pallor (97%). Most common cause of splenomegaly was hemolytic anemia (80.64%) among which thalasemia was 50% followed by sickle cell anemia 30.64%. Out of 124 patients, 123 (99.1%) received medical treatment while only one patient (0.9%) underwent surgical treatment. Among medically treated patients 18 (14.5%) were recovered completely while 100 (80.6%) improved and 4 (3.2%) stable and two patients were (1.6%) died.
Conclusions: In patient with grade III, IV, and V of splenomegaly is more likely to have hemolytic anemia as common etiology and hematological investigation should be given more emphasis in a case of splenomegaly.

Keywords: Hacketts classification, Hepatomegaly, Hemolytic anemia, Splenomegaly

INTRODUCTION

Splenomegaly is defined as enlargement of the spleen, measured by size or weight.[1] In past, splenomegaly was a clinical finding, but in recent years, imaging studies have also helped to assess for or confirm mild splenomegaly. The spleen is a functionally diverse organ with active roles in immune-surveillance and hematopoiesis.[2] The normal spleen is usually not palpable, although it can sometimes be palpated in adolescents and individuals with a slender build. However, an enlarged or palpable spleen is not necessarily of clinical significance.[3] Palpable spleen is usually accompanied by hepatomegaly and other signs and symptoms of systemic illnesses.[4] Rarely a palpable spleen may be present in a child and generally it is first suspected upon physical examination and it is defined as a palpable splenic edge felt >2 cm below the left costal margin. A palpable spleen tip may be a normal finding in up to 30 percent of neonates, the frequency drops to approximately 10 percent in healthy school-age children and <3 percent in young adults.[5]

A pathologically enlarged spleen is often firm, may have an abnormal surface, and is frequently associated with signs and symptoms of the underlying diseases.[6,7] The differential diagnosis can logically be subdivided into infectious, hematologic, metabolic, vascular, and neoplastic diseases which result in abnormalities of the lymphoid, reticulo-endothelial, or vascular components of the spleen. Splenic enlargement increases the risk of traumatic rupture of the spleen. Splenectomy, although indicated in some conditions, does not always relieve the hypersplenic state, and its benefit must be weighed against the hazard of life-threatening episodes of sepsis.[8]

The present study was an attempt to find out the frequency of various causes of splenomegaly, to study its hematological parameters and to find out role of these parameters as a diagnostic or additional tool in elucidating etiopathogenesis of splenomegaly in a tertiary care centre.

METHODS

After obtaining written informed consent from the parents/guardians, this cross-sectional observational study was conducted in 124 children of age between 0-12 years, presenting with splenomegaly and who were admitted to the pediatric ward and NICU at tertiary care center during January 2015 to June 2016. Exclusion criteria were refusal of parents or patients to participate in the study. A detailed history, thorough clinical examination, hematological and all relevant investigations were done for every child. Chest X-ray, liver function tests, Mp card, dengue serology, sickling test, HPLC (High performance liquid chromato graphy), Montoux test, Widal test, HIV, bone marrow examination, X-ray skull, blood group, blood culture, ultrasonography of abdomen, CT chest/abdomen were done when required.

The enlargement of the spleen (splenomegaly) was graded into six Grades as per Hacketts classification as given below:

- 0- Spleen not palpable even on deep inspiration.
- I- Spleen palpable below costal margin, usually on deep inspiration
- II- Spleen palpable, but not beyond a horizontal line halfway between the costal margin and umbilicus, measured in a line dropped vertically from the left nipple.
- III- Spleen palpable more than halfway to umbilicus, but not below a line horizontally running through it.
- IV- Palpable below umbilicus but not below a horizontal line halfway between umbilicus and pubic symphysis.
- V- Extending lower than class IV.[9]

Also grading of splenomegaly was done according to conventional classification as given in Hutchison's clinical methods as shown below.[10]

- Mild splenomegaly- 1-3 cm below left costal margin
- Moderate splenomegaly- >3-7 cm below left costal margin
- Massive (Gross) splenomegaly- >7 cm below left costal margin.

The prevalence and possible cause of splenomegaly in children were studied. Outcome of the study group was evaluated as regard to the complete recovery, improvement, mortality.

Collected data was entered in MS-Excel 2007 and corrected for typographic errors and analyzed using SPSS 16.0 version. The comparison of qualitative data was done using chi-square test. The confidence limit for significance was fixed at 95% level with p-value <0.05.

RESULTS

There were 8,491 admission in pediatric ward and NICU during study period amongst those 124 were found to have splenomegaly. Hence, the prevalence of splenomegaly was 1.46%. Majority of patients 42.7% were in age group of 6-12 years followed by 3-6 years (32.2%). There was male predominance with male to female ratio of 1.3:1 (Table 1).

Table 1: Age and sex distribution of the study group.

Age group	Male	Female	Total (n=124)	%
0-1 month	2	1	3	2.4
>1 m-1yr	3	7	10	8
>1y-3yr	7	11	18	14.5
>3 -6 yr	22	18	40	32.2
>6 -12 yr	36	17	53	42.7
Total	70 (56.45%)	54 (43.55%)	124	100

Fever was the most common presenting symptom found in 75% patients, followed by cough 64.5%. Family history of anemia was observed in 28 (22.5%) patients as shown in Table 2. Pallor was the most common sign found in 97% of patients followed by hepatomegaly 90% and abdominal distension 72.6%. Association of hepatomegaly with splenomegaly as clinical sign was found to be statistically significant (p= 0.03) as shown in Table 2.

Most common grade of splenomegaly according to conventional method was mild splenomegaly present in 46 patients (37.09%), followed by moderate splenomegaly in 41 patients (33.06%), and massive splenomegaly in 37 patients (29.83%). Most common grade of splenomegaly was grade III of Hackett's

classification found in 33% of the patients, affecting mainly in the children of age group 3 to 6 years (Table 3). The most common associated finding with Hackett's classification of splenomegaly was hepatomegaly found in 90% patients.

Table 2: Distribution of frequency of symptoms and clinical signs.

Symptoms	Male	Female	Total	%	P value
Fever	52	41	93	75	-
Cough	45	35	80	64.51	-
Jaundice	22	17	39	31.45	-
Abdominal Pain	22	12	34	27.41	-
Family history of anemia	17	11	28	22.58	-
Bone/joint pain	18	9	27	21.8	-
Skin rash	3	4	7	5.64	-
Bleeding manifestations	6	1	7	5.64	-
Clinical signs	**Male**	**Female**	**Total**	**%**	**P value**
Pallor	68	52	120	97	0.718
Hepatomegaly	59	52	111	90	0.03
Abdominal distension	48	42	90	72.6	0.195
Icterus	22	17	39	31.45	0.67
Lymphadenopathy	15	14	29	23.38	0.31
Edema feet	8	5	13	10.48	0.696
Ascitis	4	2	6	5	0.605

Table 3: Splenomegaly as per Hackett's classification.

Grade of splenomegaly	Male	Female	Total	%
Grade I	10	6	16	13
Grade II	17	13	30	24
Grade III	21	20	41	33
Grade IV	13	12	25	20
Grade V	9	3	12	10
Total	70	54	124	100

From the Table 4, it was observed that etiology of splenomegaly is more of hematological condition as the age advances, it is followed by infectious causes and this association was statically significant (p=0.004) (Table 4).

Most common cause of splenomegaly was hemolytic anemia found in 80.64% among which thalasemia was 50% followed by sickle cell anemia 30.64%. In hematological conditions having splenomegaly, Grade III and IV were more common and statistically significant (p= 0.0029) as compare to other causes of splenomegaly as shown in Table 5.

Table 4: Relation of age and etiology in study group.

Age	Infectious	Hematological	Congestive	Storage Disorder	Total
0-1 month	3 (2.41%)	0	0	0	3 (2.41%)
> 1 m-1 yr	4 (3.22%)	5 (4%)	1 (0.80%)	0	10 (8.06%)
>1y-3yr	2 (1.61%)	16 (12.9%)	0	0	18 (14.51%)
>3yr-6 yr	4 (3.22%)	36 (29.03%)	0	0	40 (32.26%)
>6yr-12yr	7 (5.64%)	44 (35.48%)	1 (0.80%)	1 (0.80%)	53 (42.76%)
Total	20 (16.10%)	101 (81.5%)	2 (1.60%)	1 (0.80%)	124 (100%)

Out of 124 patients, 123 (99.1%) received medical treatment according to the cause of splenomegaly while only one patient (0.9%) underwent surgical treatment (splenectomy). Among the patients who were treated medically, 46 had a mild, 41 had moderate and 37 patients had a massive splenomegaly. The only one

patient, who had undergone surgical treatment, had a massive splenomegaly. Out of 124 patients 18 (14.5%) recovered completely, 100 (80.6%) were improved and 4 (3.2%) remained same and two patients (1.6%) died.

Table 5: Etiology of splenomegaly according to Hackett's classification.

Etiology	Grade I	Grade II	Grade III	Grade IV	Grade V	Total
Thalesemia	2	8	26	17	9	62 (50%)
Sickle cell disease	4	10	13	8	3	38 (30.64%)
Malaria	3	3	2			8 (6.51%)
Liver diseases	2	2				4 (3.22%)
Sepsis	1	3				4 (3.22%)
Cardiac diseases	2					2 (1.61%)
Nutritional anemia	1					1 (0.8%)
Dengue fever		1				1 (0.8%)
Galactosemia		1				1 (0.8%)
HIV		1				1 (0.8%)
Typhoid fever		1				1 (0.8%)
Tuberculosis	1					1 (0.8%)

DISCUSSION

The prevalence of splenomegaly in present study was 1.46% thus indicating that amongst every 100 patients admitted in ward one is likely to have splenomegaly. This result was correlated with the study done by Timite et al, in which they reported 1.6% prevalence of splenomegaly.[11] The age of patients ranged from 0 months to 12 years, 57.2% children were below the age of 6 years and 42.7% cases were between the ages of 6 to 12 years. Male predominance observed in the study with Male: female ratio of 1.3:1, this was comparable with the Tmite et al, Pore SN et al, and Chandanwale SS et al, studies.[11-13]

The most common presenting symptoms were fever followed by cough and most common sign was pallor which was similar to the prior studies.[11,12,14-16] The association of hepatomegaly with splenomegaly as clinical sign was found to be statically significant. Abdominal distension was found in 72.6% patients is was because of hepato-splenomegaly which was present in most of the patients, because 80.6% of patients were having either Thalasemia or sickle cell disease. Most common grades were Hackett's grade II and III which is comparable with study done by Timite et al, where it was found in (61.8%) cases.[11] According to conventional classification, most common grade was mild followed by moderate and then massive. Similar result was found in study done by Agarwal et al.[14]

General investigations included Hb, WBC, platelets count, peripheral blood smear, CBC blood film for malaria, urine and stool examination were done in all patients. According to the clinical evaluation and the results of the general investigations a second line or specific investigations were done. These included sickling test, LFT, HPLC. Among all investigations haematological investigations played a major role in the diagnosis among which HPLC was the key investigation. The other important investigations were biochemical, serological and radiological which were helpful in the diagnosis. In present study, it was found that the grade of splenomegaly correlated significantly with the Hb level which is statically significant (p value <0.001). Hb >10 g/dl was found in 3.2% patient all of these patients have grade I splenomegaly while Hb < 10 g/dl was found in 96.8% patients of grade II, III, IV. Normal WBC and platelet count were found in most of the patients while peripheral blood picture was found to be normal in 15.3% cases. Abnormality in the RBCs shape and size was noticed among 75 patients (60.5%) while 30 patients (24.2%) had abnormality in the morphology of the RBCs.

The most common cause of splenomegaly was hemolytic anemia, this diagnosis included conditions such as sickle cell disease, thalassemia. Malaria was found in 8 cases, 2 cases had *P. falciparum* infestation and six of P. vivax. Majority of cases had Grade I-II splenomegaly. Splenomegaly in malaria was due to diffuse histiocytic proliferation, which was comparable with Pore et al, study.[12] Infectious etiology other than malaria was found in 12 cases. Other common etiologies were hepatitis (4), sepsis (4), dengue fever (1), enteric fever, tuberculosis and HIV in one case each. All these cases had grade I-II splenomegaly. Splenomegaly in these cases was due to cellular response of the spleen to circulating infectious agents and toxins. This etiological observation is comparable with Pore et al.[12] Regarding the outcome of the patients in this study was only two patients with massive splenomegaly were died while rest were discharged after treatment. Hence mortality in present

study was (1.6%). From the observations of present study, it can be concluded that in the patient with higher grade (III, IV and V) of splenomegaly are more likely have hemolytic anemia as common etiology, and hematological investigation should be given more emphasis in a case of splenomegaly.

Since thalassemia and sickle cell anemia/disease were the most common cause of splenomegaly in this area. It is to recommend that awareness of the parents that splenomegaly can occur as a consequence of various causes, with variation in the presenting complaints, should be increased through sessions of health education at the referred clinic and ward. The awareness of all treating doctors should be raised about the importance of proper clinical evaluation and proper haematological work up for any patient with palpable spleen to rule out serious causes. All hospitals need a proper setup of investigations that can confirm the causes of splenomegaly such as HPLC. Certain protocol for investigating and management of children with splenomegaly must be provided to all pediatrics hospitals vaccine, *H. infleunzae* and pneumococcal, Hep. B should be made available, accessible and affordable for those children who has sickle cell disease and thalasemia.

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